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The Forms and Differential Diagnosis of Bulbar Poliomyelitis

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Historical Introduction

References to conditions which may have been poliomyelitis dates from earliest times but no description of this disease has been found in medical literature before the end of the eighteenth century. At this time mention of acute paralysis in childhood was made in England by Michael Underwood in his textbook on diseases of children. During the subsequent years there were several descriptions of poliomyelitis, the best being that of the German orthopedist Heine (4), which was published in 1840. During most of the nineteenth century, the disease was regarded as a sporadic affliction of young infants. Its contagious and epidemic character dit not receive attention until Medins work appeared in Sweden in 1890. From that time forward the disease ceased to be a curiosity and became a periodic scourge in some countries. The principles of its epidemiology where first reviewed by Medins pupil Wickman (14) who published his monograph in 1908, the same year in which Landsteiner discovered the virus which has been accepted as the etiological agent for the disease. Three years later Kling (5) and his associates in Stockhol applied this discovery to the clinical investigation of poliomyelitis. Since that time widespread interest in the disease has lead to numerous publications on the subject. Investigation in all parts of the world has resulted in the recognition of the clinical picture of the Bulbar form of this disease.

Etiology and Transmission

Since Landsteiner, in 1909 (6) attributed the virus as being the etiological factor in poliomyelitis many investigators have confirmed his results. There is little doubt today of this. Theiler and Bauer (11) have measured the virus to be 10—17 mu. It has proved very difficult to grow in culture, however it has been successfully cultivated in the presence of nerve tissue taken from human embryos. It is very susceptible to heat, being destroyed in 30 minutes by exposure to a temperature of 45°—50° C. It is killed rapidly by ultraviolet light. On the other hand, it is resistant to freezing, drying, X-rays, sonic vibration, an numerous protoplasmic poisons. It survives in water for a month in light and 3—4 months in dark. It remains active in feces at ice-box temperature for several months, but dies out in a few days in sewage. It withstands 1 % phenol and 15 % ether for several months at 0° C. In 50 % glycerol it can be preserved for 4 to 6 years. Little is yet known about its antigenic structure, but cross-neutralization tests carried out with the sera of convalescent human beings and monkeys-show that several immunilogically distinct types of virus exist.

The mode and route of infection is still not clear. There is a mass of evidence pointing in different directions — one to the respiratory, the other to the alimentary route. The interpretation of the evidence is diffi-

cult. The evidence pointing th the alimentary route of infection is briefly that the disease occurs in the summer and early autemn months; that the virus seems to be commoner in the alimentary tract than in the nose; that it is often present in sewage; and that it has been isolated from flies. In addition, monkeys can be infected by feeding; and outbreaks of milkborne infection have been discribed. On the other hand, the disease does not behave epidemilogically like a milk-borne disease; and milk-borne infections are not uncommon in diseases such as scarlet fever and diphtheria, that are normally transmitted by droplets. Burnet showed that cynomolgus monkeys could be infected by simple swabbing of the pharyngeal mucosa with infected material.

On the whole, the evidence points toward the tonsillo-pharyngeal mucosa as being on of the commonest sites of invasion and to infection being air-borne rather than food-borne; but whether the infective air-borne material consists of infected droplets expelled from the nose and mouth or of dust contaminated with feces is still open to question. Insect-borne infection is improbable, mainly because the virus has never been demonstrated in the bloodstream, though flies might, of course, become contaminated with feces and sewage (Topley & Wilson) (12).

Classification

The term bulbar poliomyelitis becomes very confusing when we examine the variety of clinical manifestations which are included under this title. It is quite obvious that the lesions causing these symptoms are not confined to the medulla oblongata but involve the entire brain stem including the pons and the midbrain. Attemps have been made to classify the Bulbar Polio in respect to the site of the lesion. A widely accepted system is that employed by the Minnesota Poliomyelitis Research Commission. The classification finally evolved to include all cases of the bulbar form was as fellows:

(1) those in which the nuclei of the cranal nerves are involved
   a) upper cranial group, and
   b) lower cranial group

(2) those in which the autonomic centers are involved
   a) respiratory center, and
   b) circulatory center

(3) those in which there is combined bulbar and encephalitic involvement
   a) diffuse encephalitic group, and
   b) focal encephalitic group

(4) those in which bulbar and upper spinal group are involved in combination.

A short explanation of each will help in understanding the classification.

(1) Granial nerve group

a) *Upper cranial* — This includes the fifth, sixth, seventh, and eighth cranial nerves as well as the ocular nerves. This produces syndromes known as polioencephalitis sup. and inf. of the old lieterature. Isolated ocular neural palsies as well as total external ophthalmoplagias have been bescribed. Pupillary disturbances including Horner's syndrome and hippus, have also been mentioned. Disturbances of mastication result from damage to the motor portion of the fifth nerve. Palsies of the fifth nerve may be unilateral or bilateral and may involve all or part of the muscles supplied by the fifth nerve. The patient may be unable to close his mouth, but in less severe cases it may be necessary to test the movement against resistance. In unilateral lesions of the fifth nerve the jaw may deviate when the patient opens his mouth.

Paralysis of the facial nerve was frequently observed. At times the entitre ditribution of the facial nerve was involved and at other times the weakness was limited to the upper or the lower part of the face. Bilateral deafness and vestibular disturbance have been reported in the literature.

b) *Lower cranial* — Involvement of the nuclei of the lower cranial nerves, particularly the tenth, affords definite threat to the life of the patient. The nerve in conjunction with the eleventh and twelvth is essential for the act of deglutition. Brahdy described the following four types of difficulty in swollowing: regurgitation through the nose, accumulation of food ou fluid in the pharynx, regurgitation of food from the esophagus, and cough due to failure to shut off the larynx.

On examination one notices nasal speech and failure of equal movement of the soft palate. Laryngal stridor may be present. With the inability to swallow there is the constant tendency toward pooling of saliva and food in the pharynx. This accumulation causes an obstruction to the airway. A further threat to the airway results from the aspiration of fluid into the larynx or from reflex spasm of the glottis. The lesion of the tenth nerve may also produce impaired innervation of the vocal cords. This may manifest itself by a warning stridor or by rapid abductor paralysis of the vocal cords with sudden asphyxia and death.

(2) Central autonomic group

a) *Respiratory center* — In many cases symptoms of involvement of the respiratory center in the medulla have come on a number of days after the onset of pareses of the cranial nerves. During the course of the illness there develops irregularities of rythm and depth of respiration. Respiration tends to be shallow, and there are prolonged intervals between inspirations. Following this, long periods of apnea occur and finally respiration ceases. During and previous to the development of respiratory symptoms these patients are extremely anxious and apprehensive. There are periods of confusion. These symptoms are frequently caused by hypoxia and may be present before the apearance of cyanosis. Ultimately the temperature and pulse rate trend upward. The blood pressure may be elevated or fall to shock levels terminally. At necropsy there is marked hemorrhagic pulmonary edema.

b) *Circulatory center* — The symptomatology which is quite typical is as follows: The patients lips are deep cherry red, and the skin has a flushed florid appearance. The pulse rate is extremely rapid ranging between 150—200. It is often irregular and at times difficult to palpate. The blood pressure is low and later becomes unobtainable. The pulse pressure is as low as 10 mm- mercury. The heart is inaudible before the respirations cease. At necropsy the lungs show severe hemorrhagic edema.

(3) Encephalitic group

a) *Diffuse encephalitic group* — These patients show great anxiety. They are kennly aware that something is wrong with them and frequently feel that they are going to die. Ideas come to them unusually rapidly and minimal stimuli produce rapid responses. In the motor sphere these patients show quivering, trembling, twitching and jerking of the facial muscles and the extremitiel. The face is flushed. All movements are rapid as a part of

the general hyperactivity. Many of the patients are unable to sleep and remain awake for several days and nights before hyperactivity subsides. In fatal cases confusion becomes prominent and often is followed by lethargy, coma and death. Prolonged somnolence is not uncommon in children. In many patients especially those in respirators, a typical toxic confusion apears one week after the onset of the illness and lasts about one week. Not all patients with encephalitic symptoms show inflammitory lesions in the hemispheres, which leads one to believe that many of these patients suffer from hypoxia of the cerebrum. Many causes may be found such as, obstruction of the air passages or failure of the peripheral or central respiratory mechanism or circulatory failure.

b) *Focal encephalitic group* — It seems probable that focal signs are due to actual inflammitory involvement of the implicated structure. Also many focal lesions were found at necropsy which remained silent. There have been reported such focal signs as aphasia, visual verbal agnosia and cerebral motor phenomena. These include myoclonic jerks as well as Grand Mal convulsions. Spastic hemiparesis has been reported.

(4) Combined Bulbar Cervicothoracic cord group

These patients require treatment in a respirator because of the severe damage to the cervical and thoracic cord, which produces failure of the diaphragm and intercostal muscles. In children this combined lesion is not nearly as critical as in adults, in whom the prognosis is very grave. The most frequent findings at necropsy aside from the nervous system, were hemorrhagic pulmonary edema, atelectasis, bronchopneumonia, pulmonary abcess and rarely massive pulmonary embolism.

The most frequently recognized of these forms of Bulbar Polio consists of involvement of the cranial nerves. However, only destruction of the tenth cranial nerve furnishes an immediate threat to the life of the patient, by producing an obstruction to the airway and subsequent asphyxia.

In a report of 21 cases, collected from the Jenner Kinderspital, Bern, Switzerland, during a severe epidemic in the summer and fall of 1954, we may examine a few of the forms.

The most constant lesion was that affecting the facial (VII) nerve. In 15 of these cases, facial paralysis was definitely elicited. In all but six of these patients, facial paralysis was the only localized lesion found. These would be classified as «Upper cranical group». In one case we saw both upper (N. VII) and lower (NN. IX, XI, XII) cranial groups as well as a diffuse encephalitic component. In another, facial paralysis was combined with damage to NN. IX and XII and the respiratory center, as well as spinal paralysis. Besides the cases mentioned, NN. IX and XII were affected in one other case in which they were combined with a spinal paralysis. The abducens (N. VI) nerve was reported in three cases. In one it was the sole lesion, in another it was combined with a respiratory center lesion and in the third it was demonstrated along with thes pinal

and encephalitic forms. Three of the twenty one cases terminated fatally — all with respiratory tractcomplications.

It is obvious from this report that the classification used by the Minnesota Poliomyelitis Research Commission is limited. Nine of the cases from the Jenner Kinderspital were a combination of at least two types and occasionally more. One such case was that of «W. L., Age. 10».

The patient was delivered spontaneously after a full term pregnancy. He first sat at age 6 months and walked at age 11 months. Prior to his present ailment he suffered occasional angina, for which a tonsillectomy was performed in 1951. Since that time until 16. 11. 1954 the child was in apparent good health.

At that time he complained of headache and had a temperature of 37.2. The next day the child ran a fever of 37.8 and two days later he vomited and complained of pain in the neck. He was also constipated. The next day his voice was distinctly nasal and he regurgitated food through his nose. On that day he was admitted to the Jenner Kinderspital.

Physical Examination:

Eight year old male in a reduced general condition, exhibited a red face and a nasal voice. On examination he was found to have a tachycardia. Nuchal rigidity, Brudzinski, Spine sign, and Kernig were all elicited. An Amoss sign was questionable.

All reflexes were easily exhibited and no peripheral pereses were found. On extending the tongue it deviated to the left and the soft palate was paretic on the left side, being able to raise only on the right. The uvula was deviated to right. The facial musclus as well as the eye muscles and sternocleido were intact. No other abnormalities were found.

Laboratory findings:

I. Liquor

a) Pressure — moderately raised
b) Color — clear and colorless
c) Pandy — positive
d) Cells — 60/3

II. Blood

a) Hb. — 112 %
b) R.B.C. — 4.8 million
c) F.I. — 1.12
d) W.B.C. — 12 000
1. Neutr. staff — 3.5 % segmented — 70.5 %
2. Eosin. — O
3. Baso. — O
4. Lympho. — 18 %

III. Urine

a) Color — yellow and cloudy
b) Naphthyl — neg.
c) Protein — trace
d) Urobilinogen — normal
e) Sugar — neg.
f) Acetone — neg.
g) Sediment — neg.

Clinical Course:

On the second day after admission, the patient felt pain in both legs. The reflexes and tonus began to sink. By the sixth day there was a definite paresis of the left leg with weakened patellar reflex. The paresis was extreme in the foot and an Achiles tendon reflex. could not be demonstrated. After two weeks the hypoglossal and palatine paralyses were still present. The patient walked with a limp, being unable to carry his body weight on the left toes. Examination showed paresis of the flexors in the hip and foot as well as the extensors of the foot. The pareses regressed somewhat and on the twentieth day the patient was released for ambulatory treatment.

Diagnosis:

Bulbar poliomyelitis (NN. IX and XII) combined with a spinal paresis.

Therapy:

Benadon T.I.D., Redoxon 500 mg., Pyramidon-Urotropin solution 4 × 1 KL. After the first week, heat and massage therapy was instituted and continued until release.

Summary:

On the day of admission the patient exhibited a nasal voice and he regurgitated food through his nose. An examination showed him to have a tachycardia, positive Brudzinski, Spine, and Kernig signs and nuchal rigidity. The tongue was paralized sence of 60/3 cells, a positive Pandy in the spinal fluid, and a moderately raised liquor pressure, the symptoms allowed the diagnosis of bulbar poliomyelitis. Two days after admission the patient felt pain in both legs. This progressed to a severe paresis and loss of reflexes in the left leg and foot by the sixth day. The hypoglossal and glossopharyngeal paralyses were still present after two weeks. The pareses regressed and on the twentieth day the patient was released for ambulatory treatment.

In still another case we may demonstrate a combined form of Bulbar Poliomyelitis.

There was nothing of medical importance in the birth and development of P.B., age 8½ years. He has had measles varicella, and pertussis.

On 21. 9. 54 the child complained of headache. He had a fever of 38.8 and that evening it sank to 37.8. He had been previously in good health.

The next day he had temperature of 38.2. and complained of pain in the head and back of the neck. He had no pareses at that time. That same day he was admitted to the Jenner Kinderspital with the provisional diagnosis of poliomyelitis. One of his brothers was being treated at that time in the Jenner Kinderspital for poliomyelitis and subsequently a third of the four children in the family was also admitted with the same diagnosis.

Physical Examination:

The eight year old male was in a good general condition, somewhat small and frail for his age, had a clear sensorium, and did not give the impression of being seriously ill. On examination the only positive findings were a positive Spine sign, negative Brudzinski, Lasegue, and Kernig, and he was unable to completely close his right eye.

Laboratory findings:

I. Liquor

a) Pressure — raised
b) Color — clear and colorless
c) Pandy — positive
d) Haine — positive
e) Cells — 724/3, 76 % Poly., 24 % Mono.

II. Blood

a) Hb. — 80 %
b) R.B.C. — 4.4 million
c) W.B.C. — 10 700
1. Neutr. staff — 0.5 % segmented — 71.0 %
2. Eosin. — O
3. Baso. — O
4. Lympho. — 26.0 %
5. Mono. — 2.5 %

III. Urine

a) Color — yellow and cloudy
b) Protein — neg.
c) Naphthyl — neg.
d) Urobilinogen — normal
e) Sugar — neg.
f) Acetone — neg.
g) Sediment — occasional leuco., oxalate, mucus.

Clinical Course:

Two days after admission the patient still ran a fever of 38.1. He exhibited a Meningismus, Kernig and Spine sign. His muscle tonus and reflexes were normal. The right facial nerve was affected, the patient not

being able to close the right eye. On the next day the child became very somnolent. The examining physician was unable to elicit answers from the child. Two days later the patient was much less somnolent and answered questions though somewhat slowly and only after repeated query. He was slightly atactic on intention. The facial paralysis become stronger and by the seventh day all three branches were affected, predominently the middle and upper. He still showed meningeal and spine. signs. On the ninth day he no longer showed the Meningeal or Spine sign and was no longer lethargic. No improvement of the facial paralysis was seen. This situation did not change for five or six days. Then the facial paralysis and general condition got progressively better and on the twenty-first day the patient was released. At that time a paralysis of the middle and upper branches was still to be seen. The child was then treated ambulatory with Galvanization and infrared therapy.

Diagnosis:

Bulbar Poliomyelitis — Upper cranial nerve (N. VII) and encephelitic involvement combined.

Therapy:

Benadon T.I.D. Redoxon 500 mg, Pyramidon-Urotropin solution 4 × 1 KL. From fifth day on, heat and massage therapy.

Summary:

The patient was admitted to the hospital in the acute stage of poliomyelitis. The diagnosis was made by the demonstration of a Spine sign, a right-sided facial paralysis, onset with headache and fever, and the presence of an epidemic. He did not exhibit any encephalitic symptoms upon entry. The liquor findings supported the diagnosis, mainly a cell of 724/3, a positive Pandy, and a raised liquor pressure. On the third day he developed encephalitic signs (somnolence and muscular hypotonus). The paralysis remained localized to the facial muscles and the encephalitic signs receded by the ninth day after admission. By the twenty first day the facial paralysis showed some improvement and the patient was released.

A third case exhibited a fulminating course in which total paralysis and death resulted.

B. S. was delivered spontaneously at term after a normal pregnancy on 11/10/45. At age 5 he had measles, at 7 years he had varicella and pertussis at 8 years.

On the morning of 8/6/54 with localized pain at Mc Burney's point. His temperature was measured rectally to be 39.7. The pain increased with a leucocytosis of 9700. An appendectomy was performed that afternoon. The appendix was only very slightly injected and a fecolith was found at the tip. No enteritis or exudation was demonstrated. That evening and next day the patient was in a good general condition. On the evening of the second day his breathing became labored and meningeal signs were elicited.

His reflexes were normal with no pareses. His pulse rose to 150 and his leucocyte count was 11 400 with 80 % seg., 2.5 % staff, 14 % lymph., 3.5 % mono., Basal pneumonia was found on the left.

On 10/6/54 his temperature was 38.4, he was cyanotic and still showed meningeal signs. His sensorium was clear. He had been getting penicillin since the operation. He was admitted to the Jenner Kinderspital on 10/6/54.

Physical Examination:

An 8 3/4 year old male entering the hospital exhibited a greyish blue cyanotic color. He breathed with difficulty and with the help of his auxillary respiration muscles, and even these moved very poorly. His eyes were opened wide, his mouth was full of mucous and he spoke with difficulty but clearly. During the examination his condition became worse and he became more cyanotic.

His heart was normal, Over his lungs was found a dullness on the left base dorsally. Over this area was heard bronchovesicular breath sounds. Meningeal signs and head-drop were positive.

The arm reflexes were increased. The patellar and achilles tendon reflexes were normal, Babinski were negative. The both upper arms were paralized, both forarms could be moved — but with difficulty. The boy could not sit up or turn. He could not swollow his saliva but could swollow tea. His pharynx was injected.

Laboratory findings:

I. Liquor

a) Pressure — raised
b) Color — cloudy
c) Pandy — positive
d) Cells — 364./3
e) Haine — sugar 105 mg %

II. Blood

a) Hb. — 107 %
b) R. B. C. — 5.3 million
c) F. I. — 1.01
d) W. B. C. — 16 600
   1. Neutro. staff — 2.5 %
      seg. — 90.5 %
   2. Eosin. — 0
   3. Baso. — 0
   4. Lympho. — 5.5 %
   5. Mono. — 0.5 %

III. Urine

a) Sugar — neg.
b) Acetone — trace

Clinical Course:

During the course of the afternoon of the day of admission the boy's voice became unclear and difficult to understand. For the past 16 hours he was unable to swallow because the fluid would be extruded through his nose. His sensorium also became cloudy. His legs, forarms and hands could still be moved but with difficulty. That evening an attempt to turn off the iron lung caused severe cyanosis. At that time, fluids (tea and Dextopur) were begun rectally. Throughout the night the patient was stimulated with Sympatol and Lobelin ½ c. c. and 1 c. c.

At 3 o'clock the next morning the child complained that he had strong pains in his legs and was unable to move his arms or legs. Immediately after that he was unable to speak. Therapy with Strychnine, Benadon and Lobelin was continued.

At 6 o'clock his condition was markedly worse with severe cyanosis appearing even with the administration of 02 and stimulants. His pulse was rapid and weak. The boy was calm.

At 8 o'clock he began to lose consciousness, corneal reflexes became weaker and the cyanosis of the lips became more pronounced. His temperature was 40.5. He was given Largactil, Irgapyrine and Coramine.

At 9 o'clock the child continued on a downhill course dispite continued therapy.

Exitus letalis came at 12 o'clock noon. There was no autopsy performed.

Diagnosis:

oCmbined bulbar and spinal poliomyelitis with a complicating pneumonia.

Therapy:

02, Strychnine 0.001, Benerva sortissimum, Lobelin 1A., Gamma Globulin 2 c. c., Blood transfusion from father, penicillin 4 × 200 000, Sympatol 4 × 0.5 c. c., Iron lung, Cibalgin, tea and Dextropur.

Summary:

B. S. was born 11/10/45 and was a healthy child with a history of common child illnesses (measles, varicella, pertussis). With localized abdominal pain and a slight leucocytosis of 9700 with no shift to the left he was operated on for appendicitis. There was no demonstration of an infected appendix. Post-operative-ly he quickly developed basal pneumonia. Both, the operation and the pneumonia obviously lowered the boy's resistance but we cannot say definitely that this accounted for such a fulminating course. It is probable that the abdominal pain was a premonitory symptom of the poliomyelitis.

The diagnosis of poliomyelitis is allowed on the basis of a meningismus, progressing total paralysis, a typical spinal fluid and history. We may classify this case as being a bulbar form on the bases of the difficulty in swallowing, regurgitation through the nose and the difficulty in speech. The

Angström Respirator was indicated here but was not used due to the extremely fulminating course. The pneumonia diagnosed by auscultation and percussion.

The patient arrived at the Jenner Kinderspital in a state of cyanosis, dyspnea, meningeal signs, and paralysis of the upper arms. In the period of one day the condition of the patient swiftly degenerated to total paralysis and unconsciousness and eventually death.

Diagnosis and differential diagnosis

In a full blown paralytic case, diagnosis is relatively easy. The characteristic history and physical findings of stiff neck and back, reflex changes and paralytic (bulbar) findings together with an abnormal spinal fluid leave little doubt.

a) *Characteristic history* — There is fairly rapid onset of fever, malaise, headache, sore throat, and sometimes vomitting, which may occasionally be the most predominent symptom. It may be coupled with abdominal pain to bring the differential diagnosis of acute appendicitis into consideration. This stage may last for one to several days, when symptoms start to dissappear, the patient becoming well. Then there follows a reoccurence of fever and hadeache accompained by C.N.S. involvement.

b) *Physical findings* — In the preparalytic stage we find meningeal signs and reflex changes. The symptoms of meningeal irritation are typically:

1. The spine sign — Because of pain and stiffness in the Lumbar region, the child is unable to kiss his own knee.

2. The Amose sign — The patient has difficulty in righting himself from a lying position. To remain in the sitting position the patient must support himself with his arms.

3. The Lasèque sign — The patient suffers pain in the ischiadicus area upon raising the outstretched leg.

4. Kernig's sign — This consists of pain and resistence on extending the leg at the knee and flexing at the hip.

In bulbar polio, the first indications may be a report by the parent that the child's voice has taken on a nasal tone, or that there is difficulty in swallowing, sometimes resulting in fluid being extruded from the nose. With progression, the inability to swallow becomes more serious and mucus and saliva accumulate in the throat. These signs may progress with dramatic rapidity, while the patients distress increases with disorientation, shock and coma.

The *cerebrospinal fluid* shows important diagnostic findings in poliomyelitis. In the acute stage of the disease it is under normal or slightly increased pressure, and a delicate fibrin clot may form on standing. The cell count, highest in the preparalytic stage and rapidly falling after onset of the paralysis, averages about 150/3 cells per cu.mm., polymorphonuclear leucocytes predominating in the first 24 hours, and lymphocytes predominating later. The protein is normal or slightly elevated early and is likely to show an increase in the first two or three weeks. Thus we have at the

beginning of the disease a «dissociation cyto-albuminique» and after the first one or two weeks a «dissociation albumino-cytologique». The normal or increased sugar content differentiates poliomyelitis from T.B. meningitis; the sugar content and the absence of bacteria, from bacterial meningitis; the negative Wassermann from syphilitic meningitis. A normal spinal fluid, particularly a normal cell count, is almost never found in the preparalytic stage of polio.

The *blood picture* in polio is not changed. There is neither a change in the white cell count or a shift to the left. The sedimentation rate is also normal or very slightly increased. Several diseases may afford difficulty in diagnosis of the preparalystic stage of bulbar polio. The confusion lies mainly in the meningeal signs found in these afflictions. A rheumatic disease comes into consideration in this stage. It may take the form of lumbago, torticollis rheumatica, or sciatic pains. It is ushered in by many of the symptoms observed in the beginning stage of polio, mainly: vomiting, fever, headache, pains in the back, neck and extremities and a positive Lasque sign. We may observe a weakness in the leg muscles which may be confusing. However we do not find a change in the reflexes or the spinal fluid. Finally we may diagnose a rheumatic disease by the successful use of antirheumatic drugs.

T.B., meningococcus, and other meningitides come into consideration in the earlier stages. There are several points of difference in both. T.B. meningitis and bulbar polio may both be accompanied by vomiting but it is usually more severe in T.B., polio begins suddenly whereas T.B. is more insidious in its onset, and course. In polio there is found an early loss of reflexes, however they may be at first hyperactive. As mentioned above, the cerebrospinal fluid is very important. In T.B. there is a reduction of the spinal fluid sugar to about 50 mg %, a large increase in cells and a clouding. The polio patient is not as stuporous as the T.B. victim. The final diagnosis can be made by exhibiting a miliary tuberculosis or miliary tubercles in the retina.

In the meningococcal meningitis we find a pussy cerebrospinal fluid with many thousand cells, mostly polymorphs with intracellular diplococci. The neck stiffness and opisthotonus are much more pronounced than they are in polio. M. meningitis usually occurs in the winter and early spring. The Kernig sign usually appears much earlier in M. meningitis, in polio appearing on the 4—5 day. There is also found a typical meningeal type of breathing where there occurs rythmically spaced apnoic periods between normal respirations. Usually M. meningitis exhibits a hemorrhagic exanthem which is not seen in polio. The blood picture is typical of bacterial infections whereas in polio it remains unchanged.

In the nonparalytic stage it is further necessary to differentiate polio from «*Schweinehüter*» disease which is a meningites caused by a spirochete. The prognosis is good as opposed to bulbar polio thus making a diagnosis important. The most important point of differentiation is the fever curve.

In Morbus Boucher — Gsell, the first fever period lasts, as a rule, 4—5 days, (by polio 1—2 days) and the temperature is higher than in the second stage. Also, very often a herpes or a conjunctivitis may accompany the disease. By Gsell a bradycardia is present and by polio a tachycardia. Also then, very important is the history of some contact with swine.

The polyradiculitis of Guillain — Barre poses an important differential diagnostic problem. It comes into consideration both in the pre- and in the paralytic stages. It usually has a shorter prodrome than does polio, the paralysis is not as severe and is symmetrical. The cerebrospinal fluid shows a «dissociation albumino-cytologique» which is the reverse of polio in the beginning. The recovery of paralysis in Guillain — Barre is more complete and more rapid than in poliomyelitis.

Important in the differential diagnosis of bulbar poliomyelitis is a post-diphtheritic paralysis. The most often affected structure is the uvula. However this paralysis and as well as any other post-diphtheric paralysis is symmetrical as opposed to bulbar poliomyelitis. Facial paralysis and paralysis of the ocular muscles and extremities also occur. Also there will be a history of diphtheria in the previous month, usually about 3 weeks before onset of the paralysis, Glanzmann (3).

Tumors of the C.N.S. present symptoms similar to those of C.N.S. disease. The diagnosis of a brain tumor is difficult in the early stages but it should not be confused with lower motor neuron disease such as poliomyelitis. Choked disks are said to be present in about 90 % of the cases, however it depends on the location of the tumor. The disease is slow in onset, except in gliomas, with progressively severe headache and dizziness, Wechsler, (13). Paralysis is spastic in type although it may be flaccid early and is usually hemiplegic. Fever is not a prominent finding. The spinal fluid pressure is elevated and the protein level is increased. Cerebral lesions may cause unilateral flaccid or hypotonic paralysis of the lower side of the face. As opposed to polio, tumors also cause sensory disturbances including astereognosis and vasomotor changes. In the cerebrospinal fluid we find all grades of pleocytosis and the protein may be as high as 4000 mg %.

Cerebellopontine angle tumors may grow from the fifth, seventh, or eighth nerves or the petrus portion of the temporal bone. Occassionally they are bilateral or are a part of von Recklinghausen's disease. Subjective noises preceeds the onset of impaired hearing. This is followed by the ipsilateral paralysis of the sixth and seventh nerves and possibly neuralgia in the region of the trigeminus. At the same time vertigo and nystagmus set in because of involvement of the vestibular nerve. Later cerebellar symptoms may make their appearance. Choked disk is a late phenomenon. The spinal fluid shows very marked increase in proteins. Aggrevation of signs may follow lumbar puncture. There may also be early vomiting, nausea, and vertigo; disturbance of equilibrium, and inclination of the head toward the lesion.

Tumors of the pons begin as a rule with focal signs, while the general symptoms follow very late. The characteristic signs consists of alternating hemiplegias and paralysis of the lateral gaze. Nystagmus especially vertical,

is pathognomic. Crossed anesthesias, that is, of the face on the side of the lesion and of the body on the opposite side is another characteristic sign.

Tumors of the medulla are rare and also give alternating hemiplegias, with cranial nerves 8—12 impaired on the side of the lesion. As the medulla is a small structure the symptoms are often bilateral. Paralysis of the vocal cords, dysarthria, atrophy of the tongue, nystagmus, ataxia, vertigo, and vomiting are often present. The tumor is rapidly fatal. General tumor symptoms (headache choked disk, convulsions, stupor, and pulse and respiratory changes) come on late or are completely absent.

Tumors of the midbrain including the cerebral peduncles and the colliculi may lead to a Weber or Benedikt syndrome. These numerous symptom complexes caused by this type of tumor are due to the vast number of nuclei and vegetative centers in the area. Tumors of the third and fourth ventricle, tegmentum, pendunculi, etc. all give different tumor symptoms.

Cerebrovascular lesions may be confused with bulbar polio due to the paralytic symptoms. The paralysis starts off flaccid and may become spastic. Furthermore, a positive Babinski or other pyramidal reflexes are exhibited in C.V.A. The sudden development of unconsciousness followed by hemiplegia, and usually cranial nerve weakness rules out poliomyelitis.

Hysteria may exhibit paralysis suspicious of polio. Differentation is not always easy. Weakness of muscle groups may be a persistent complaint. Reflexes, however are normal to hyperactive. There is sensory disturbance but it is bizarre in nature. There is usually no meningeal signs or spinal fluid changes. A careful history can usually illuminate the emotional basis.

Relation of Symptoms to Lesions

The Pons:

Because of the frequent presence of facial paralysis in bulbar polio, it was apparent to the early investigators that the pons was commonly involved. Wickman (14) was one of the first to describe the pontine form of poliomyelitis. During the Swedish epidemic of 1905 he collected 26 cases of facial paralysis, 6 cases of abducens paralysis, and 2 cases of trigeminal paralysis. It was apparent to these investigators that three types of facial paralysis occuued in this disease: (1) facial paralysis appearing as an isolated finding (2) facial paralysis appearing with paralysis. The first of these, namely the isolated facial paralysis is of utmost importance because it indicates that at times the pons may be the most severely involved of all the structures of the brain stem.

Except for the involvement of the seventh cranial nerve nucleus in poliomyelitis, reports indicating injury to other pontine structures are few indeed. Because of the proximity of the sixth cranial nerve to that of the seventh, it is not surprising that this nuclei group is the second commonest pontine structure reported involved in polio, Wickman (14). Only scattered reports suggest damage to other pontine structures, such as deafness, trigeminal paralysis, vertigo.

Since many patients with bulbar polio are too ill to be examined carefully, the clinical observations are often inaccurate and do not present the complete picture of the severity of the tissue involvement. Moreover, many of the pontine structures are difficult to examine clinically in a very ill patient. It is for this reason that one is forced to resort to pathologic studies to determine the severity and extent of pontine involvement in this disease. As early as 1894, Redlich pointed out that all the pontine structures, the lateral vestibular nucleus (Deiter) and the acoustic nucleus were the most severely damaged in bulbar polio. These observations have been repeatedly verified.

It is interesting to note that, although clinically the facial nucleus seems to be the most prominently involved in pontine poliomyelitis, pathological studies reveal severer damage to some of the other pontine structures. Many of the structures are silent clinically and it is likely that pontine polio is commoner than is generally realized.

In a recent study, Matzke and Baker (9) reported on 109 cases of bulbar poliomyelitis. Involvement of the cranial nerve nuclei in the pons was observed in 49 of their cases. Fifth nerve impairment, manifested by disturbance in the muscles of mastication occured in twelve cases. In three cases trismus was noted. The complication appeared suddenly, usually at the end of the first week of illness, and persisted from five to seven days. In somepatients the involvement of the muscles of mastication is not detected until months have elapsed, at which time, difficulty in chewing or actual atrophy of muscles with facial assymetry first become apparent. Paralysis or paresis of the facial muscles (seventh nerve) was observed in 33 of their cases. Disturbances of the eye muscles (fourth and sixth nerves) are extremely difficult to evaluate, however, histologically these nuclei were moderately to severely involved in almost half the cases; nevertheless clinical impairment is recorded in a much smaller percentage of the cases. Histologic studies showed the pons to be involved in practically every case. This involvement consists of both a diffuse interstitial cell infiltration and neuronal damage. The degree of histologic damage far exceeded the amount of clinical dysfunction observed.

Irritation of the fifth cranial nerve nucleus may result in severe trismus. The upper half of the vasomotor center is situated in the large reticular cells of the lower part of the pons. Involvement of this region of the pons can produce rapid circulatory collapse and death in the absence of any medullary disturbances.

The Hypothalamus

Due to the tremendous progress in the treatment of polio many more patients are returning home, apparently cured. However, many of the symptoms of hypothalamic involvement do not manifest themselves until the patient leaves the protected and comfortable environment of the hospital, and is exposed to the stresses of every day life. A serious occasional case is that in which the patient suddenly succombs weeks or months after an apparently complete recovery, and no specific cause can be discovered

for this tragic outcome. The work of many authors on the diencephalon in bulbar polio has aided in clarifying the situation. It has long been known that the nuclear groups within the hypothalamus play an important role in the maintaining of the internal environment of the human organism and in preserving the stability of its visceral, somatic, and even emotional functions. Dysfunction or irritation of the hypothalamus has been reported as resulting in disturbance of blood pressure, gastrointestinal carbohydrate metabolism, fat metabolism, regulation of body temperature, and even emotional responses. In most cases the predominent involvement was observed in the walls of the third ventricle, with implication of the paraventricular nuclei.

In a study of 115 cases of bulbar poliomyelitis by Baker, (8) lesions were found in 85 % of the cases in the hypothalamus. This high percentage has been out by other investigators. However, many of the manifestations are not detected clinically since many of the tests for hypothalamic activity are complicated and require the aid of the patient. The most striking changes seen were the (1) alteration in loss and conservation of heat, manifested by hyperthermia and hypothermia: (2) disturbances in vascular control, manifested by the acute onset of hypertension: (3) variations in sleep and wakefulness: (4) disturbance in the regulation of gastrointestinal function as indicated by acute gastric hemorrhage or gastric stasis.

The following signs of disturbance in temperature regulation due to hypothalamic involvement in bulbar poliomyelitis suffice for a diagnosis: (1) An acute onset of very high fever after a period of moderate hyperthermia: (2) persistance of hyperthermia throughout the illness in spite of the subsidence of other clinical evidence of the acute illness: and (3) absence of any evidence of pulmonary or urinary disease process, Rammon (10).

Hypothermia is much less common than hyperthermia. Many patients in spite of very severe poliomyelitis, often complicated by extensive pulmonary involvement, showed normal or only slightly elevated temperature. As a result of recent investigations it it is generally believed that the anterior portion of the hypothalamus is concerned with the mechanism of heat loss and that lesions in this area will result in hyperthermia.

In many patients severe hypertension develops during the course of the illness but it is difficult to evaluate this as being a hypothalamic or a medullary disturbance. The symptom is usually transient lasting 3—4 days.

Damage to the anterior portion of the hypothalamus, involving the tuberal region, results in gastric hemorrhage and erosion. This region of the thalamus is concerned with parasympathetic activity, and its destruction cancan be expected to result in impaired digestion and inhibition of gastrointestinal peristalsis, with marked distention. Very often gastric hemorrhage is witnessed and evidenced by vomiting of gross blood, failing blood pressure, and a rising pulse rate. In milder forms we find «coffee ground» material in vomitus aspirated from the stomach.

Months or even years after the recovery from bulbar poliomyelitis patients will show evidence of subclinical damage to the hypothalamus as evidenced by their abnormal response to special tests used to determine hypothalamic function.

Mesencephalon

The midbrain has received but scanty attention in various publications on bulbar poliomyelitis. In a number of cases of bulbar poliomyelitis one can be certain that the midbrain has been involved because of the presence of ocular signs, Emmons (2). It is estimated that some ocular signs will be apparent in about 10 % of cases of bulbar poliomyelitis even without careful neurological examination. As early as 1905 Wickman (14) reported such involvement in 56 of 868 cases of poliomyelitis. In most cases pathological investigation demonstrated lesions within the colliculi and the third nerve nuclei.

Matzke and Paker (7) in a very thorough study of 111 cases of patients who died of bulbar poliomyelitis found mild to severe changes in the substantia nigra in 31 %. They also detected mild changes in the trochlear nuclei and severe damage of the oculomotor nuclei in 37 % of the cases. This damage implicated more than 50 % of the cells. The substantia nigra and the tegmental nuclei were constantly hit. However symptoms are much less severe than might be expected when these extrapyramidal nuclei are affected.

From the study of the literature it seems that the midbrain is never as severely implicated than the lower regions of the brain stem. Also, within the midbrain the severest involvement is generally observed in the oculomotor nuclei, the substantia nigra and the tegmental nuclei. The lack of correlation of symptoms to the lesion can possibly be explained by the rapidity and severity of the disease, not allowing time for the detection of extrapyramidal symptoms. Occasionally, post-poliomyelitis parkinsonism has been reported in those patients who survive.

Summary

Bulbar poliomyelitis is a term frequently used inaccurately. The word bulbar would tend to connote the medulla oblongata. It is the purpose of this dissertation to clarify the designation of the various forms and the lesions involved, causing them. The lesions in bulbar poliomyelitis are found from the cerebrum to the medulla oblongata, including the pons mesencephalon, and hypothalamus. The most frequently affected structure is the pons. This was confirmed by the figures at the Jenner Spital which showed that of the 21 reported cases, 15 manifested facial paralysis which may be attributed to a lesion in the pons. Two of these cases are discussed here and demonstrate that a combined form exists. Such combined forms were common in the epidemic in Bern in 1954. A system of classification of the various forms was evolved by the Minnesota Poliomyelitis Research Commission and seems to be the most sensible. This system is used in this dissertation.

A differential diagnosis of bulbar poliomyelitis is extremely important since the prognosis and treatment differs from many disorders that present a similar clinical picture. As mentioned, in the nonparalytic stage we differentiate bulbar poliomyelitis from tuberculous and meningicoccal meningitis, rheumatic diseases, appendicitis, Morbus Boucher — Gsell and polyradiculitis of Guillain-Barre. Polyradiculitis also comes into consideration in the paralytic stage along with post-diphtheric paralysis and tumors of the central nervous system.

One of the major advances in the treatment of the bulbar form of poliomyelitis is the use of the Angström Respirator combined with a tracheotomy. Until recent times it was the fate of these unfortunate patients to die of asphyxiation in an iron lung due to the closing of the larynx by mucous masses. As soon as difficulty with swollowing sets in and interferes with respiration due to the collection of secretions in the upper respiratory tract, a tracheotomy should be performed a canula inserted and the glottis closed. The patient should be placed in an Angström Respirator.

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